Seizure 32 (2015) 72–74

Contents lists available at ScienceDirect

Seizure

journal homepage: www.elsevier.com/locate/yseiz

Clinical letter

Jeavons syndrome as an occipital cortex initiated generalized epilepsy: Further evidence from a patient with a photic-induced occipital seizure

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ARTICLE INFO

Article history:
Received 4 August 2015
Received in revised form 5 September 2015
Accepted 8 September 2015

1. Introduction

Jeavons syndrome (JS) is characterized by the triad of eyelid myoclonia with or without absences, eye closure-induced generalized paroxysms and photosensitivity. Occasional generalized tonic–clonic seizures may also occur. Despite having been claimed as a distinct entity among the IGEs by several authors, its inclusion as an epilepsy syndrome in the ILAE classifications is still a matter of debate. The findings of both interictal and ictal posterior focal abnormalities during eyelid myoclonia have led to the hypothesis of considering JS as an occipital cortex initiated generalized epilepsy [1]. Moreover, a recent study using EEG-fMRI and voxel brain morphometry protocols in patients with JS demonstrated anatomo-functional abnormalities involving a circuit that includes the occipital cortex and the cortical/subcortical systems physiologically involved in the motor control of eye closure and eye movements [2].

Here we describe a patient with JS who presented a photic-induced focal seizure with electroclinical semiology typical of the occipital lobe. To our knowledge, focal seizures of occipital origin have not been previously reported in patients with JS.

2. Case report

A 24-year-old female was first seen in our department at the age of 19 years for evaluation of uncontrolled, sporadic, generalized tonic-clonic seizures (GTCS) despite being on antiepileptic drug treatment. An uncle had a history of a single GTCS at age 15 years. At the age of six years, she presented with a GTCS during a febrile illness. An EEG showed a single generalized spike-wave discharge during hyperventilation. Treatment was not started and she remained asymptomatic until the age of 18 years. At this age she presented an unwitnessed episode of loss of consciousness while using her computer. A previous feeling of dizziness was recalled by the patient. A brain MR was normal and an EEG showed several, spontaneous and hyperventilation-induced, generalized irregular polyspike and spike-wave discharges. Treatment with valproic acid, lamotrigine and topiramate failed because of adverse events. She was finally started on levetiracetam 1500 mg daily.

A third, apparently spontaneous, GTCS occurred at age 19 years. Her mother, who witnessed the episode, described repetitive blinking before the fit. A video-EEG recording showed normal background activity and frequent discharges of generalized polyspike-waves, most occurring immediately after eye closure (Fig. 1a) and commonly accompanied by eyelid myoclonia. Consciousness impairment could not be demonstrated despite an exhaustive investigation. Other relevant EEG findings were unilateral focal sharp waves in both temporo-occipital regions during sleep (Fig. 1b) and a generalized photoparoxysmal response.

A diagnosis of Jeavons syndrome was made. Her relatives then recognized that EM considered “motor tics” were present since childhood.

Levetiracetam was switched to zonisamide 300 mg daily and a control video-EEG was performed 3 months later. Photic stimulation at 12 Hz induced a generalized photoparoxysmal response immediately followed by a focal seizure with occipito-temporal ictal onset (Fig. 2). Clinically the first symptoms were bilateral asymmetric blinking, predominant on the left side, and intermittent cephalic and conjugate eye deviation to the left, followed after several minutes by visual agnosia, unresponsiveness, forced

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http://dx.doi.org/10.1016/j.seizure.2015.09.003

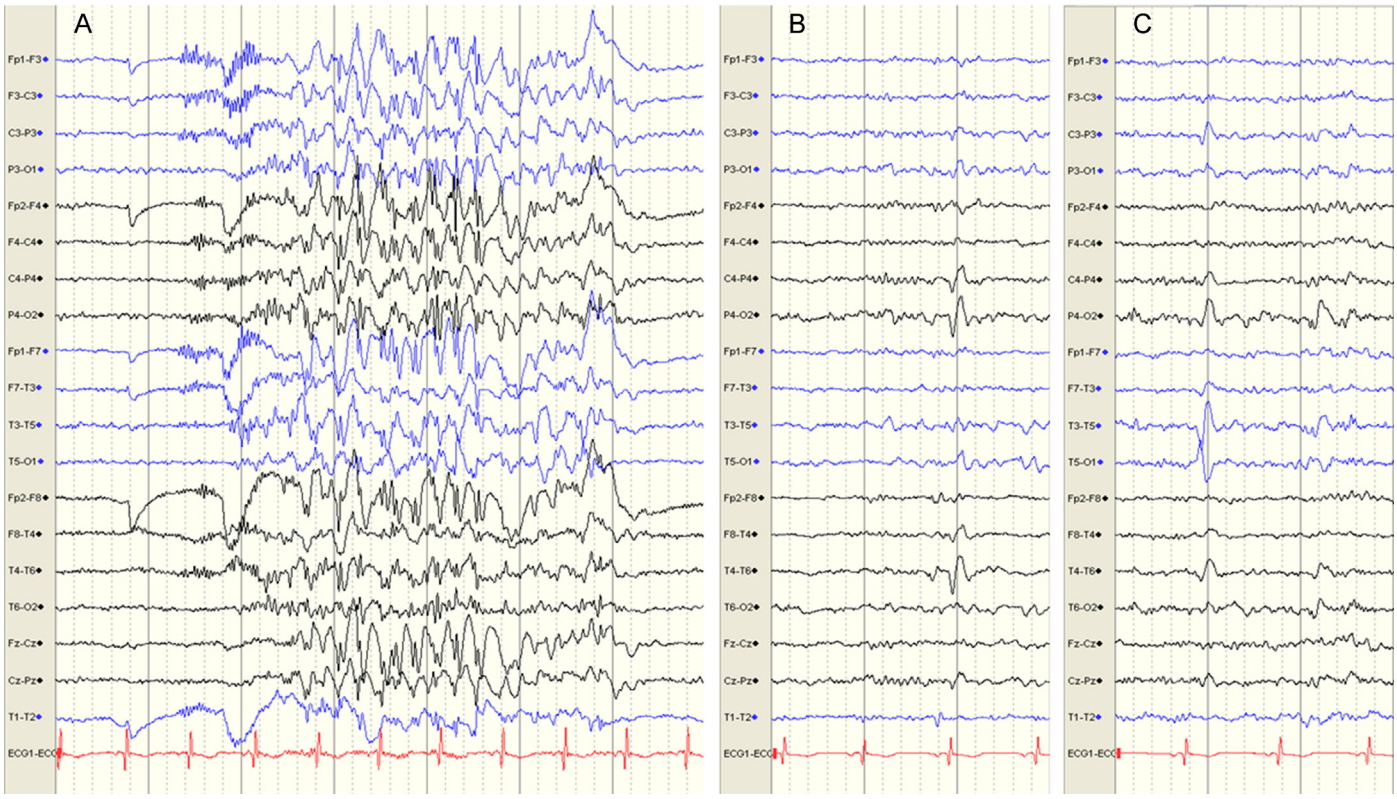

Fig. 1. Following eye closure, a brief (0.40 s) bilateral spiky alpha activity and intermixed occipital spikes precede generalized, high amplitude, irregular polyspikes and spike-waves associated with eyelid myoclonia (A). During sleep focal *sharp* waves over both occipito-temporal regions were frequently recorded (B and C).

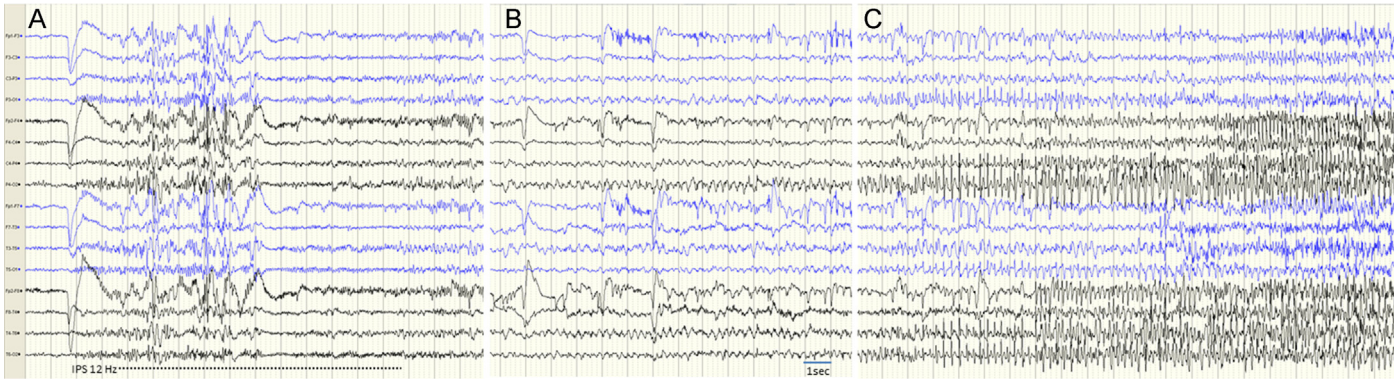

Fig. 2. Occipital lobe seizure induced by IPS at a frequency of 12 Hz. Immediately after a generalized photoparoxysmal response a low voltage fast activity appears over both occipito-temporal regions (A) which progressively evolves predominantly in the right hemisphere increasing in amplitude and decreasing in frequency (B). After 2 min from the onset it is followed by a rhythmic spike-and-wave discharge in the right occipito-temporal region that propagates to the left homologous area (C) before generalization (not shown).

oculocephalic deviation to the left and tonic-clonic convulsions. Blindness was apparent during the first 30 s in the postictal state.

Valproate was added and zonisamide later withdrawn. Since then the patient has remained free of GTCS for five years, although eyelid myoclonia still persists.

3. Discussion

In more than half of patients with photic-induced seizures subjective symptoms lead to a suspicion of an ictal onset in the occipital neocortex. However, focal seizures precipitated by intermittent photic stimulation (IPS) are considered to be a rare form of epileptic photosensitivity. In these cases, the ictal discharge originates in the occipital cortex and may remain localized or spread to adjacent areas [3]. The majority of IPS-induced focal seizures have been reported in patients with partial epilepsy (idiopathic or symptomatic) with or without spontaneous occipital seizures [3,4]. Anecdotal cases of photogenic focal seizures have been reported in patients with idiopathic generalized epilepsy [4].

The patient with JS here presented unarguably presents an IPS-induced focal seizure originating in the occipital lobe, as the ictal EEG shows. Generalized epileptiform discharges and eye-closure sensitivity have been described in patients with occipital partial epilepsy, mainly in idiopathic photosensitive occipital epilepsy (IPOE) [3]. Our patient, however, has never presented the typical

seizures recorded in patients with IPOE which are characterized by elementary visual symptoms followed by epigastric discomfort and vomiting with or without impairment of consciousness.

According with Viravan et al. [1], in JS specific clinical and electroencephalographic features would raise the possibility of the occipital cortex initiating a generalized epilepsy network including thalamocortical and transcortical pathways and the brainstem. Additionally, the IPS-induced focal seizure recorded in our patient would support the role of the visual cortex as a "seizure generator" in this epileptic syndrome. The recognition of a specific cortical area, in the context of diffuse cortical hyperexcitability, is not uncommon in other idiopathic epilepsies. In this view, JS might be probably better categorized as "system epilepsy" rather than as a generalized or focal syndrome.

Conflict of interest statement

The authors declare that there are no conflicts of interest.

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